CASE REPORTS

PAROXYSMAL ATRIAL FLUTTER IN PERONEAL MUSCULAR ATROPHY

BY

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Cardiac involvement is known to occur in several of the heredo-degenerative disorders of the nervous system; for example, it is recognized in Friedreich's ataxia (Evans and Wright, 1942), dystrophia myotonica (De Wind and Jones, 1950), and in the muscular dystrophies (Manning and Cropp, 1958). However, no reference to cardiac involvement in peroneal muscular atrophy (Charcot-Marie-Tooth disease) could be traced. In this case report, paroxysmal atrial flutter and cardiac failure occurred in a patient with peroneal muscular atrophy.

Case Report

A 59-year-old steel worker first began to complain of short episodes of nausea and dizziness two years ago. These attacks, which were of sudden onset, were always accompanied by a regular "beating" in the epigastrium, and on one occasion by polyuria. They gradually became more frequent and latterly were associated with breathlessness. During the week following a particularly severe attack, which lasted several hours, he felt increasingly tired, became progressively more breathless, and noticed swelling of the ankle for the first time. In the past, he had enjoyed good health and his only illnesses were an injury of the left wrist thirty years ago, which resulted in a traumatic ulnar palsy, and an attack of right-sided sciatica seven years ago which had improved following treatment in a plaster jacket.

On examination, he was slightly dyspnœic. The heart rate of 100 beats a minute was regular in time and force. The blood pressure was 115/70. The heart was not enlarged and there were no murmurs. There was evidence of cardiac failure in the presence of engorged neck veins, hepatic enlargement, and bilateral basal crepitations.

In addition, the patient had wasting of the small muscles of both hands, and in the legs wasting of all the muscle groups below the knee, giving him a drop foot gait and bilateral pes cavus. There was loss of vibration sense in both feet, but all other forms of sensation were normal, apart from some anæsthesia in the ulnar distribution of the left hand. Both ankle jerks were absent, but all the other reflexes were within normal limits. There was no nystagmus or ataxia. The pupils were of normal size and shape, and reacted normally to light and on accommodation. The neurological abnormalities, therefore, suggested a diagnosis of peroneal muscular atrophy, and this was confirmed by electromyography (Dr. J. A. Simpson). The thyroid was not enlarged and there was no clinical evidence of thyrotoxicosis.

An electrocardiogram and a two-step exercise tolerance test had been taken a year before the onset of cardiac failure, and showed minor T wave abnormalities in the left ventricular surface leads, but no evidence of myocardial ischæmia. Other investigations carried out at this time included a chest X-ray, which showed the cardiac outline to be normal and the cardiothoracic ratio 15:33, and a four-hour ^{131}I uptake of 12 per cent. The serum cholesterol was 210 mg. per 100 ml., and the Wasserman and Kahn reactions were both negative. A further cardiogram taken at the time of the development of congestive cardiac failure (Fig. 1) showed flattening of the T wave in aVL, V5, and V6, as the only abnormality.

The day after this record was taken, the patient complained of an uneasy feeling in the stomach, accompanied by a fluttering sensation and increasing breathlessness. On examination, the heart rate was 140 a minute; it was regular, but there was pulsus alternans. The electrocardiogram showed atrial flutter (Fig. 2). The patient was given digitalis, the ventricular rate slowed, and normal sinus rhythm was restored five days later. The cardiac failure cleared rapidly and he was able to return to work. Although he continues to

take digitalis, he has had several more attacks of palpitation but these are less disabling than formerly. There has been no significant change in his electrocardiogram.

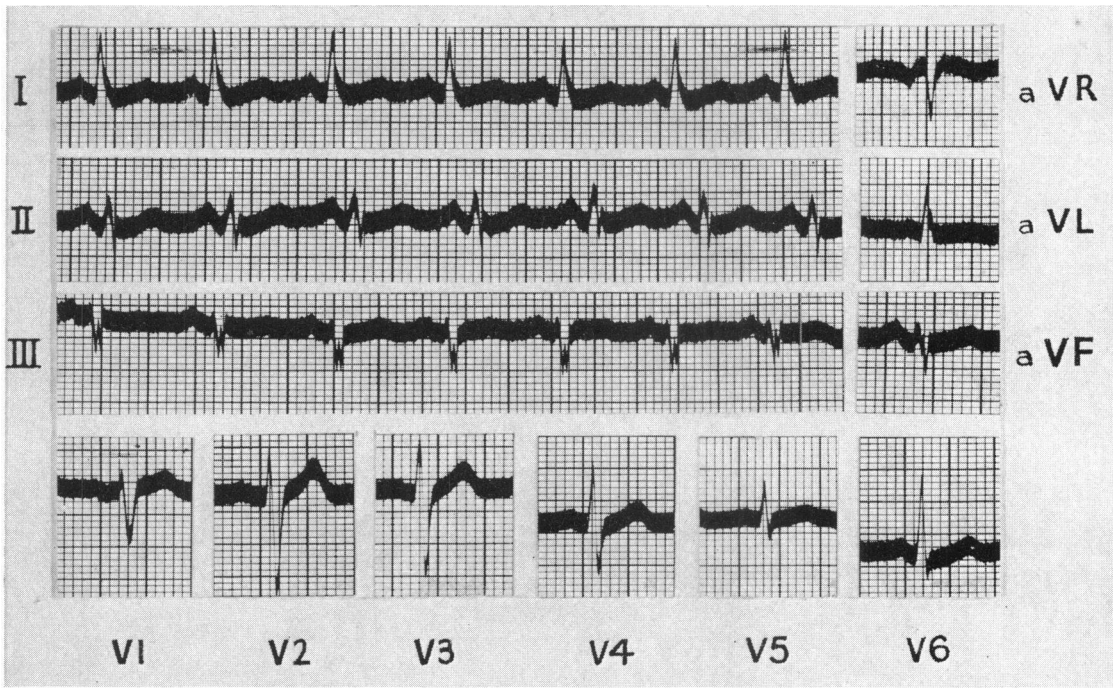

FIG. 1.—Electrocardiogram taken shortly after the development of cardiac failure in a patient with peroneal muscular atrophy. There is flattening of the T wave in aVL, V5, and V6.

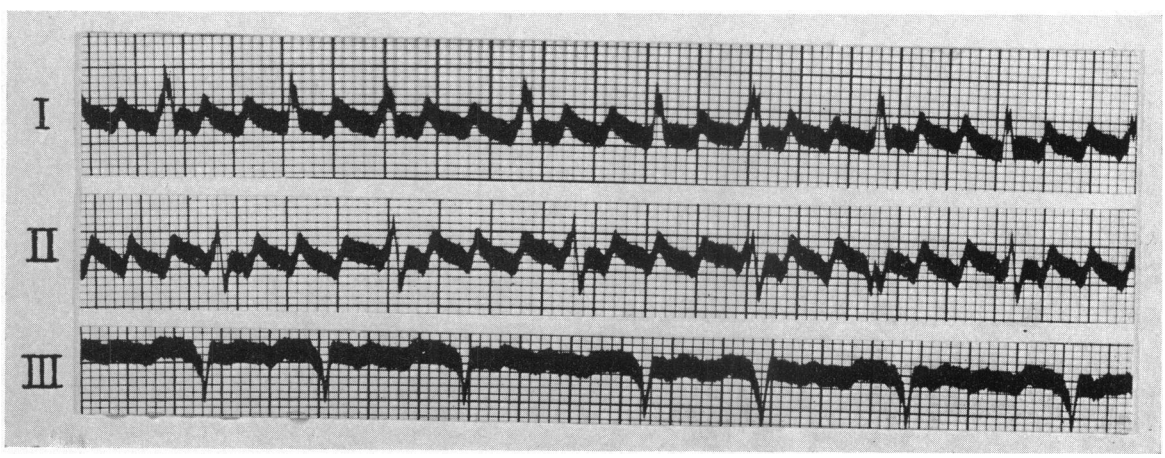

FIG. 2.—Electrocardiogram showing atrial flutter, with varying A-V block.

Discussion

From the evidence available in this case, there is little to suggest a diagnosis of ischæmic, hypertensive, rheumatic, or thyrotoxic heart disease. Wood (1956) has pointed out that atrial flutter is rare in otherwise normal subjects, and the question whether it could be associated with peroneal muscular atrophy must be considered. Friedreich's ataxia is genetically related to peroneal muscular atrophy (Hierons, 1956). Isolated instances of Friedreich's ataxia occur in families with peroneal muscular atrophy (Biemond, 1928), and combined forms of the two diseases in the same subject have been described (Ross, 1942). In view of this close relationship, it seems reasonable to suppose that visceral disturbances may also occur in peroneal muscular atrophy, and this has been supported by the recent report of gastro-intestinal manifestations of the disease (Norstrand and Margulies, 1958).

The cardiac abnormalities that are recognized in Friedreich's ataxia include flattening and inversion of the T waves in leads reflecting the left ventricular surface (Evans and Wright, 1942), and the occurrence of paroxysmal arrhythmias, occasionally complicated by congestive cardiac failure (Russell, 1946). It will be appreciated that these abnormalities are very similar to those occurring in the present case and, in view of the close relationship between the two disorders it seems probable that cardiac involvement may be an occasional complication of peroneal muscular atrophy.

Summary

Reference is made to the absence of reports of cardiac involvement in peroneal muscular atrophy. A case of peroneal muscular atrophy is described in which paroxysmal atrial flutter and cardiac failure occurred. Reasons are advanced for suggesting that the cardiac arrhythmia may be directly related to peroneal muscular atrophy.

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